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2 **A Loeys-Dietz Patient with a Trans-Atlantic Odyssey**

3 **Repeated Aortic Root Surgery ending with a Huge Left Main Coronary Aneurysm**

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Summary

We present a patient who required several aortic root procedures. A huge aneurysm of the left main coronary artery required emergency surgery after failed transcatheter procedure.

Introduction

Repeated aortic root surgery may be a challenging procedure. In case of prosthetic material infection, complete resection of the foreign material is mandatory in addition to adequate antibiotic treatment. Aortic homograft is a good option to control hemostasis and the infectious process. This Loeys-Dietz patient presented with a pseudoaneurysm of the left main following multiple root procedures.

The 25 year-old patient presented initially with aortic root aneurysm. Because of a very asymmetric pseudobicuspid aortic valve (Sievers type I, subcategory L-R) and the strong wish of the parents to avoid anticoagulation, he received a tissue valved conduit in Bern in 2003 (Shelhigh Inc. NJ, USA – presently BioIntegral, Canada). Recovery was uneventful but echocardiography at 6 months surprisingly demonstrated a limited dehiscence at the proximal anastomosis. Re-exploration showed that the suture line was not healed. Infection was excluded in presence of normal clinical and laboratory findings and unsuspecting intraoperative inspection. A pericardial patch was used to reinforce the suture. Evolution was favourable.

In 2008, he presented with a degenerated tissue valve requiring reoperation. The procedure was performed at Johns Hopkins University. The bio-conduit was resected and a mechanical composite graft implantated. The day after, the patient underwent repair of pectus excavatum. Recovery was uneventful.

Yearly echocardiographies and/or MRI were normal. In 2014, he developed fever and pneumonia was diagnosed. Blood cultures remained negative but staphylococcus was revealed on PCR. Several weeks later, echocardiography and MRI showed a partially thrombosed aneurysm of the left main coronary artery (Figure 1). PET-scan was not conclusive. The parents were against a 4th operative procedure and asked several opinion-leaders for a transcatheter technique. A covered stent was recommended to exclude the aneurysm. However, stable positioning of the stent between the ostium and the distal left main was not successful and obstruction of the left main occurred requiring resuscitation. Percutaneous ECMO was installed. The patient was immediately moved to the operation theater. Echocardiography showed a LV-EF of 10-15%).

Re-sternotomy was performed and ECMO switched to cardiopulmonary bypass through central cannulation. Intraoperative inspection showed thrombotic material inside the graft. To expose the aneurysm, pulmonary artery was transected above the valve, thrombotic material was removed. Continuity of the left main was restored with a short vein graft. Aortic homograft was implanted in the mini-root technique and pulmonary artery re-anastomosed during reperfusion. ECMO support was necessary during 48 hours. The patient was discharged after 5 weeks with compromised LV-function (EF 25-30%).

Antibiotics were continued for 6 weeks although intraoperative cultures remained negative. 18 months postoperatively, LV-EF has stabilized at 40%. The patient is doing well and findings are normal (Figure 2).

Comment

Repeated aortic root surgery is a surgical challenge (1). Aneurysm of the left main coronary artery is infrequent but may occur in patients with connective tissue disease or as a result of infection.

This patient's history presents several critical decision-making points:

- Initial biological composite graft was a wrong decision. A mechanical composite-graft would have been the best long-term option since in 2003, a valve sparing procedure was not an established standard in young patients with connective tissue disease and pseudobicuspid valve. The parents were strictly against anticoagulation.
- Dehiscence of the suture observed early postoperatively is a problem that we and other have described following implantation of this conduit (2,3).
- Etiology of the coronary aneurysm was uncertain. PET-scan was not conclusive and intraoperative cultures remained negative.
- Transcatheter attempt to exclude the left main aneurysm was not a good decision. Sudden coronary obstruction lead to resuscitation and required immediate ECMO support.
- Transection of the main pulmonary artery greatly facilitated exposure of the left main.

97 Surgery was challenging because re-sternotomy was performed under resuscitation and
98 after repaired pectus excavatum. Aortic homograft was found the best option to control
99 intraoperative hemostasis (1). Re-attachment of the coronary arteries required short
100 vein graft interposition to avoid tension and exclude the aneurysm.

101 We conclude that indication for surgery should never be weakened because
102 transcatheter intervention seems theoretically possible. In this case, catheter
103 intervention turned the repair into an emergency situation and compromised
104 significantly the myocardial function. In addition, decision-making was challenged by
105 the opinion of non-expert relatives instead of a decision based on experience and
106 expertise on technical adequacy.

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124 **Figures**

125 **Figure 1**

126 A. (Pseudo)-aneurysm of the left main artery on conventional CT-scan (A) and 3D
127 reconstruction (B)

128 **Figure 2**

129 Postoperative 3-D reconstruction of the aortic homograft with separate interposition of
130 two short vein grafts for the left and right coronary arteries